Left atrial myxoma with embolization presenting as an acute infrarenal aortic occlusion

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We present the case of a 42-year-old woman who had acute total infrarenal aortic occlusion resulting from embolic implantation from a left atrial myxoma. We propose that the small aortic aneurysm that was discovered and repaired in this case may be a direct result of invasion and destruction of the aortic elastic laminae by implanted myxomatous tissue originating in the primary atrial tumor. This behavior has been noted in small vessels of the cerebrum and upper extremities with this lesion, but no prior reports of this occurrence in the aorta has been noted after extensive review of the literature. (J Vasc Surg 1997;26:341-5.)

We present the case of a 42-year-old woman who came to the emergency department with acute onset of back pain and lower extremity weakness followed by paralysis. Total infrarenal aortic occlusion was noted on an aortogram. The cause of this sudden presentation was subsequently diagnosed as a left atrial myxoma with embolism to and occlusion of the infrarenal aorta.

CASE REPORT

The patient was a 42-year-old woman who had sudden back pain and lower extremity weakness and paresthesia, quickly followed by paraplegia, while driving to work. On admission, the patient was found to have absent bilateral dorsalis pedis pulses on physical examination and absent signals by Doppler examination. All lower extremity dermatomes from L2 distally demonstrated dense paresthesia; although there was no initial bowel or bladder dysfunction, the patient was unable to void urine spontaneously after removal of her Foley catheter. The patient was transferred to a tertiary care facility for further evaluation. On admission, an aortogram was obtained and revealed a tapering flow defect with an infrarenal aortic occlusion, consistent with either a thrombus or dissecting aneurysm (Fig. 1).

The patient was taken emergently to the operating room for exploratory laparotomy. Attention was focused on the aorta, and as the retroperitoneum was entered extreme care was taken not to disturb the collateral circulation. External examination of the aorta revealed a fusiform, expanded segment extending from the superior mesenteric arterial origin to the level of the renal arteries, with a greatest circumference of approximately 3.5 cm. It was noted that pulsation terminated at the level of the renal arteries. The decision was made at this time to clamp the aorta to approach this occlusion from above the superior mesenteric artery. The left renal artery, the right renal artery, and the superior mesenteric artery were sequentially dissected and controlled with vessel loops. The iliac arteries were cross-clamped, and an aortic clamp was then placed above the level of the superior mesenteric artery and beneath the celiac artery. The aorta was then transected and the aortotomy extended distally to the level of the inferior mesenteric artery, at which point the mass appeared to terminate. On opening the aorta in this region, a hemorrhagic, partly gelatinous mass was discovered that extended to the level of the renal arteries. This mass was firmly adherent to the intimal surface, and a prominent amount of thrombus was noted to extend from this lesion in the direction of blood flow, with extension into and occlusion of the bilateral iliac arteries. Obstructing thrombus was cleared and the affected 7 cm length of aorta was excised with the mass in situ and was retrieved en bloc. Extreme care was taken to avoid embolization to the visceral vessels. The aortic clamp was then moved below the renal arteries, and flow as restored to the visceral vessels. The total suprarenal clamp time was less than 10 minutes. The aorta was then repaired with an end-to-end anastomosis using an interposition 14 mm Hemashield Dacron graft.

Grossly, the lesion consisted of a 7 cm portion of aortic wall with an attached 3.5 cm gray-tan gelatinous mass with adherent abundant fresh thrombus. Microscopically, the lesion consisted of eosinophilic proteinaceous material consistent with organizing fibrin, surrounding numerous cleft-like, pseudovascular spaces that were not lined by

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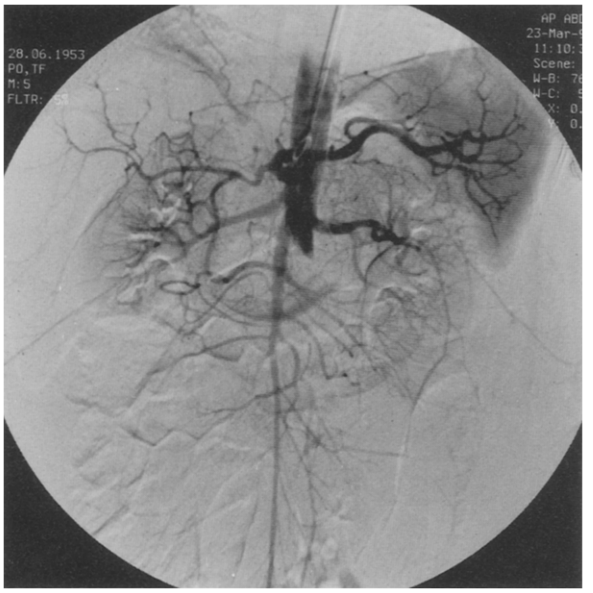

Fig. 1. Aortogram displays tapering flow defect adjacent to renal arterial orifices. Note complete absence of flow distal to the obstruction.

endothelium. Surrounding these areas were prominent accumulations of basophilic to amphophilic myxoid material, which contained a population of scattered, mildly pleomorphic giant cells that displayed abundant, pale eosinophilic cytoplasm and poorly defined cell borders (Fig. 2).

A transesophageal echocardiogram performed 3 days after the aortic operation revealed a 2 × 2 cm left atrial mass that was attached to the free wall of the chamber and extended anteriorly, focally involving the interatrial septum. The patient was returned to the operating room 5 days after her abdominal aortic operation for removal of the left atrial mass. Under cardiopulmonary bypass, the heart was arrested with cardioplegia. Left and right atriotomies were performed along the interatrial groove. The lesion was found to attach to the junction between the septum and the right free wall of the left atrium, anterior to the orifice of the right inferior pulmonary vein. The tumor did not involve the mitral valve or extend into the left ventricle. Total excision of the tumor, including portions of the septum and left atrial wall where the tumor attached, was performed. By incorporating the defect into the left atriotomy suture line, we were able to achieve satisfactory repair of the left atrium. The patient was weaned from cardiopulmonary bypass without inotropic support after 35 minutes of ischemic time and 51 minutes total on bypass.

The specimen consisted of a 2 cm portion of gelatinous, focally calcified tan-pink material. Microscopically, the lesion was practically identical in every respect to the material previously removed from the aortic aneurysm, displaying scattered groups of round, plump, and occasional stellate cells interspersed within an abundant myxoid matrix (Fig. 3). Activated histiocytes were also noted diffusely within and around the groups of mesenchymal cells, which is characteristic of this lesion. At the point of attachment of the lesion to the atrial wall, focal destruction and invasion of cardiac muscle was noted (Fig. 4). Areas of true ossification, calcification, and peripheral fibrosis suggested that the lesion was of long standing.

In light of the gross and microscopic similarities between the lesions removed from the aortic aneurysm and the left atrium and the firmly adherent nature of the aortic lesion, embolism and implantation of atrial myxoma in the abdominal aorta is considered the most likely scenario; the sudden lower extremity paraplegia was a result of the subsequent formation of a large perimyxomatous thrombus that occluded the aortic lumen.

The patient was subsequently discharged to a rehabilitation facility 6 days after undergoing atrial myxoma resection. At the time of this report, 10 months after her operation, she reports some focal improvement in sensation in the lower extremities; however, her paralysis is irreversible.

DISCUSSION

Primary cardiac neoplasms are uncommon and are usually an incidental finding in as many as 0.28% of adult autopsies.[1,2] Of these, atrial myxoma is the most common, accounting for approximately 50% of all primary cardiac tumors. Seventy-five percent of myxomas arise in the left atrium.[3] Although benign lesions, they can by virtue of their location in the blood stream produce significant morbidity in the form of vascular occlusion or embolism; as many as half of these patients present with focal neurologic deficits.[4]

Clinically, these lesions can present with obstructive symptoms such as syncope, dyspnea, and transient cyanosis as a result of positional obstruction of the mitral valve outflow tract. Palpitations and cardiac arrhythmias may also be noted, and congestive heart failure as a result of prolonged outflow compromise may be a contributing or primary cause of death in these patients.[5]

During physical examination, cardiac auscultation of these individuals can disclose the distinctive "tumor plop" shortly following S2; this sound can diminish or vanish altogether with changes in patient position.[6] The use of transesophageal echocardiography is now gaining widespread acceptance as the most useful method for a timely diagnosis.[7]

The therapy of choice for atrial myxomas is surgical resection. Atriotomy is performed, followed by visualization, careful palpation, and total resection of the tumor while avoiding the generation of embolic fragments. When the lesion involves the interatrial septum or valve leaflets, care must be taken to avoid damage or perforation of these structures. Valve re-

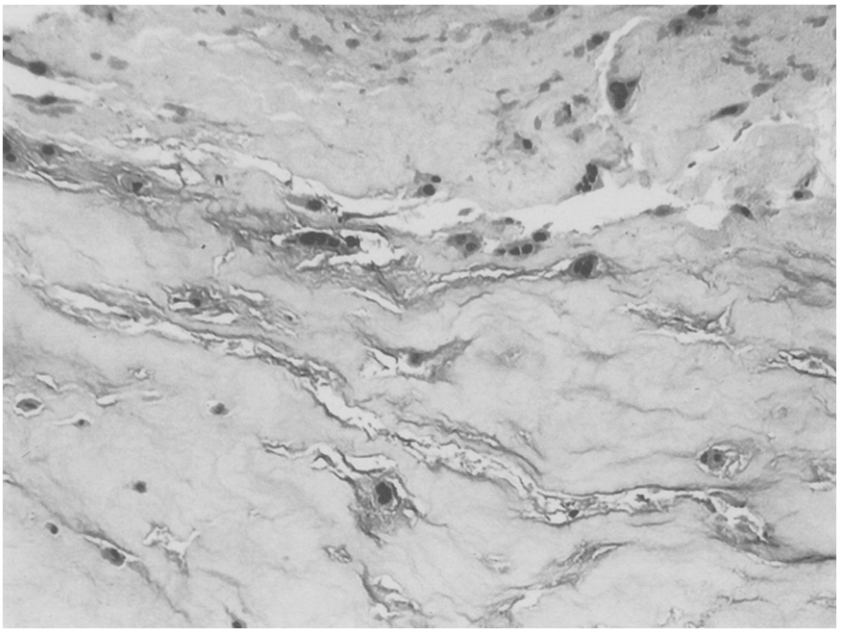

Fig. 2. Microscopic view of material removed from aorta in the area of obstruction. Note prominent accumulations of acellular fibrinoid material surrounded by areas of basophilic myxoid change and scattered large cells showing mild pleomorphism and poorly defined cell boundaries (hematoxylin and eosin; original magnification, 200×).

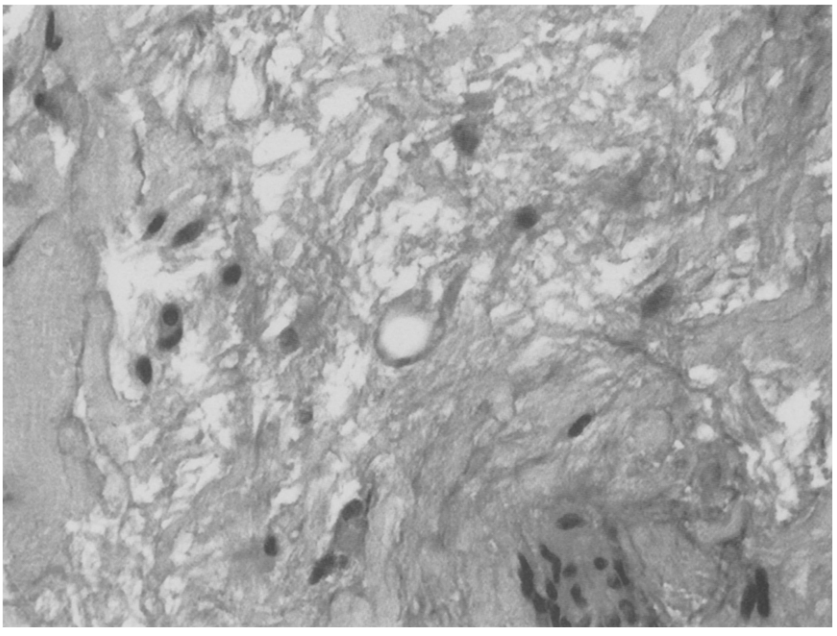

Fig. 3. High power microscopic view of resected atrial myxoma. Note similarity to the aortic lesion; mildly pleomorphic cells are embedded in a myxoid and fibrinoid stroma (hematoxylin and eosin; original magnification, 400×).

placement, in particular, has been associated with an increased postoperative mortality risk in these patients.[8] Our approach to the resection of the left atrial lesion from our patient is outlined above.

Despite the commonly friable nature of these neoplasms, careful excision can produce an intact specimen. Grossly, the lesions are often gelatinous, well-circumscribed, polypoid growths attached to a

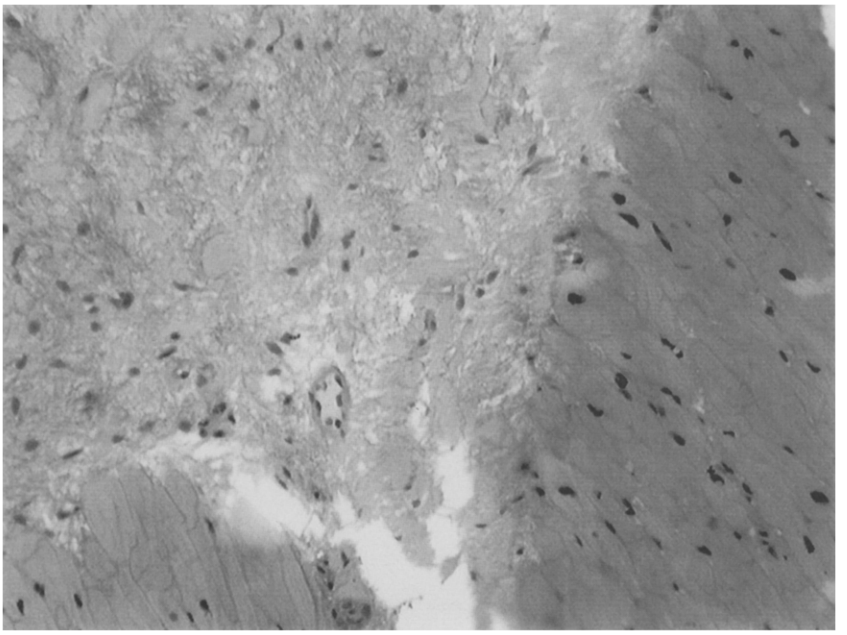

Fig. 4. Microscopic view of resected atrial myxoma. Note here the adherence to and superficial invasion by the tumor of underlying cardiac muscle (hematoxylin and eosin; original magnification, 200×).

stalk of varying width. Serial sectioning of the masses commonly reveal fibrous septae and more solid fibrous areas. Histologically, the lesions consist of plump, stellate to round cells that are associated with thin-walled vessels and an abundant basophilic myxoid matrix.

Embolization of cardiac myxoma is a common sequela (reported in as many as 30% in one series[9]). Cases in which embolism and syncope occur have been associated with a high risk of sudden death.[10] Therefore, it is prudent to consider cardiac myxoma in any young patient who has symptoms indicative of systemic vascular occlusion, and any embolectomy specimen should be carefully examined for gross and histologic evidence of possible myxomatous character.[9]

Of particular interest in this case, atrial myxomas, spreading either by systemic embolization or by true hematogenous metastasis, have been associated with the formation of aneurysmal dilatation of involved arteries a result of elastic tissue damage from tumor adherence to and invasion of the vascular wall.[11] Another potential complication of this type of tumor growth involves bacterial colonization of the primary myxoma in septic patients, which can give rise to septic emboli and abscess formation at distant sites; in addition to the inherent difficulty in eradicating these foci, bacterial infection of weakened sections of arterial walls can accelerate aneurysmal formation or rupture of involved vessels.[5] In fact, the finding of arterial aneurysms at various sites on an angiogram obtained for another cause may be the initial finding that leads to the ultimate diagnosis of cardiac myxoma.[5,12]

One could speculate that the small abdominal aortic aneurysm discovered in this case may have arisen as a result of the embolic implantation of atrial myxoma fragments and subsequent damage to the elastic laminae of the aortic wall. The presence of ossification in the primary lesion in this case indicates that the tumor is of long standing, and an embolic implant to the aorta would have had ample time to expand and invade the vascular intima to disrupt the deeper layers. This behavior is certainly possible, given the documentation of numerous examples of myxoma-derived aneurysms within the cranial vessels and arteries of the extremities.[11,13] Of course, because the lesions in our case were diagnosed practically simultaneously, it is difficult to definitively characterize the long-term behavior of the lesion, and any explanation of the origin of the aortic aneurysm must remain purely speculative.

CONCLUSION

We present a case of a young woman with sudden onset of lower extremity weakness as a result of embolic occlusion of the juxtarenal aorta. Subse-

quent investigation revealed a left atrial myxoma as the source of the obstructive material within the aortic lumen and may have served as the ultimate cause of the small aortic aneurysm that necessitated graft repair. Review of the literature uncovered no prior reports of an abdominal aortic aneurysm associated with the embolism of an atrial myxoma.

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